Paul Nabil Bustany Fund
for synovial sarcoma research
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PNB FUND NEWSLETTER

Winter 2018

In this issue…

Dear Friends,

On a beautiful October day in Morristown, in 2016, we remembered Paul Nabil, on the tenth anniversary of his death, together with Board Members and fifty friends of this organization. On this occasion we invited Dr. Kevin B. Jones of the Huntsman Cancer Institute at the University of Utah to give a live video talk about his research on synovial sarcoma and the progress made these ten years and we thank him for his excellent presentation and for his commitment to this very special research, which he continues, supported in part by this Fund. We have asked Dr. Jones again for a new update for this Newsletter which you will find later in this issue.

Recently, we were delighted to learn that this year, the National Institute of Health (NIH) awarded The Big Grant for Cancer Research to Dr. Jones. This award is only given to those on the cutting edge of their science and is recognition of his diligent work and contribution to synovial sarcoma research. The Paul Nabil Bustany Fund has been supporting Dr. Jones' research for the last nine years. Dr. Jones will be working in co-operation with two other giants in this field, Dr. Mark Ladanyi at MSKCC in New York, and Dr. Thor Nielsen at the University of Vancouver, Canada. Their collaboration will be focusing among other subjects on: *Epigenetic Mechanisms Driving Synovial Sarcoma Genesis.* We strongly believe that collaboration among the experts will not only lead to earlier results on this particular cancer but also open doors for other cancers to follow.

In the last two years we have been awarding grants to Dr. Jones on a new quest to identify and evaluate crucial genes required for synovial sarcoma metastatic spread, for example, to the lungs. This research is ongoing and in 2017 the PNB Fund awarded Dr. Jones a research grant of $50,000 and now, in 2018, he and his team will receive a research grant of $40,000.

On behalf of the Board, we appreciate all the wonderful people who have made donations to this work. In addition, it is important to mention that all of our work is done by volunteers, and we'd like to acknowledge the important services provided by our Board Members as well as our Research Proposal Advisor Unnur Thorgeirsson, MD. We are indebted to these individuals without whom we would not be able to donate a substantial research grant annually.

With gratitude and appreciation,
Laufey Bustany

The Paul Nabil Bustany Fund for Synovial Sarcoma Research Board of Directors: Rami Badawy, Christine Bustany, Laufey Bustany, Samir Ted Bustany, Alex Kaplan, Asu Okyay, Alex Rafal, and Sabrina Tom

In Memorium: Elodie Espesset (1972-2018)

Elodie was a friend of this organization and a synovial sarcoma patient, whom many of you may remember from our events over the years. My last message from Elodie was on February 2, 2018. She wrote: *"Hi Laufey, I am getting a pneumonectomy on Monday. They will remove my entire left lung. They did a lot of tests to make sure I could live with whatever is left on the right side. I hope it will buy me more time."*

Elodie was operated on February 5, and she passed away the next morning. She had already had five operations on her left lung and three on the right before this last one. She had also lost her left leg, which was the beginning of her ordeal with this deadly disease, synovial sarcoma, fifteen years earlier.

Elodie was originally from France, but lived in Edison NJ, when I first met her, together with her husband, Oleg Ayupov, her son, Alandis, and her mother, Odile, in 2008. She had seen our website and called to get some information and I invited them to my home. From that time on we became friends and stayed in touch and shared new information about the disease whenever one of us found something noteworthy. Elodie was very smart, an optoelectronic engineer, who kept on top of the field of synovial sarcoma by reading and attending sarcoma meetings, and became so well informed that she was asked to review new synovial sarcoma grant proposals of another organization.

As a moderator on the Synovial Sarcoma Survivors Network—an online community for survivors—Elodie was relentless in providing support to new patients and those having a recurrence. She had a great sense of humor, was outgoing, and approached life with a big smile.

Elodie was fortunate to have a wonderful and supportive family when she was confronted with this incurable cancer. In a piece about her mother becoming a caretaker, published by The Liddy Shriver Sarcoma Initiative, she wrote: ”*After my diagnosis, my mom crossed the ocean and the border on her own, and became our shopper, cook, cleaning lady, my 4-month-old son’s babysitter, my driver and my nurse*”—all without knowing a word of English.

An interview with Elodie, from our 2013 Newsletter, is republished later in this issue.

It is with deep sadness that I say good-bye to Elodie, my friend of ten years, who stood strong and faced her fate with exceptional generosity of heart and courage.

Laufey Bustany
December 2018

Interview: Elodie Espesset on Synovial Sarcoma and Finding a Cure

This interview originally appeared in the December 2013 Newsletter. At the time, Elodie had been a survivor of synovial sarcoma for ten years. We spoke over the phone the day after Thanksgiving (she at home in New Jersey, I across the country) and given the holiday season, our conversation naturally revolved around her friends and family, especially her husband, Oleg, and her son, Alandis. But Elodie also spoke at length about the challenges she faced with the healthcare system. It was in part because of her personal experience that she became active on the Synovial Sarcoma Survivors Network. She wanted to share her knowledge with other people going through this disease, to help them make medical decisions without regret. She felt close to her online community and enjoyed their friendship—a look through the site at the countless thank you notes she received and it’s obvious that the feeling was mutual. Elodie’s generosity and kindness, her honesty and humor, transcended distance and inspired all who knew her.

Interview by Sabrina Tom

What is the Synovial Sarcoma Survivors’ Network?
It’s a community. It’s a place where you can meet other people with the same disease. You can start discussions. You can blog. You can share information about yourself. It’s empowering.

You’re very active on the site. You’re always wishing people happy birthday and asking about their families.
Well I’m trying to find out what’s going on with them. I think it’s very useful to know what happens to other people. I cannot foresee my future. At one point I may have to make some decisions about my health and to know what’s happened to other people can help me make those decisions.

You’re also an advocate for seeing a synovial sarcoma specialist.
I think it’s important. It’s a rare disease and if you don’t take care of it right away the right way, that can make a big difference.

Is this coming from personal experience?
Yes, and from stories I've heard from other people. Some people regret not having been more aggressive with the disease at the beginning stage and then the disease comes back and they feel they didn't do everything right. I don't want people to regret anything. I think they should see someone who is familiar with the disease, not someone who's only seen one case.

What's your relationship to this online community?
I feel a lot of empathy for people because I'm going through the same thing so I feel close in that way.

That says a lot about your spirit.
I don't know how to express it. When I talk about my disease to my friends—my real friends that I see everyday—I don't feel comfortable. I feel they feel sorry for me. But for these people who have this same disease I feel more free to talk about it.

You have a common language.
Yes, exactly.

What's been your experience with the healthcare system?
Pretty awful. In France it's government-run health care. Everything is virtually free. You never have to worry about it. When I was diagnosed here I had to worry about it. For example, where they sent me first for my biopsy, the hospital was not covered by my insurance. So I tried to find a different surgeon who would be covered by my insurance and I wasn't able to do so. My insurance wasn't covering any sarcoma surgeons that I could find on the internet. I asked my doctor to refer me to a sarcoma surgeon and the only one he knew was at the Newark hospital and that hospital wasn't covered by my insurance. So I asked more surgeons, "Do you know any other guys?," and they didn't want to tell me any other names.

So what did you do?
I did the biopsy with the surgeon and then once I got the diagnosis I decided to get a different opinion from an oncologist who was not in my insurance, either. But I was very impressed with him and his hospital was covered so I decided to go with him. I did have to pay out of my pocket his fees. I was lucky in the sense that my work had short-term insurance so even though I wasn't working during the treatments I was receiving 60% of my salary. So that helped me pay for it. Without that it would've been difficult because my husband and I had just bought a house, just had a baby, so maybe it was the worse time really.

What would you like say to our community about this disease?
I do hope they find a cure quickly. We really need some new treatment. It can get depressing when I look at all the people I've met that have passed away. And people are young. Sometimes they have young kids. It's really sad.

There are so many cancers and this doesn't seem to be one that gets attention.
When something is rare, it's not interesting for the big pharmaceuticals. How do you get the funding to go forward with the research, all the clinical trials and everything?

It's important to keep being an advocate and show that there are a lot of people fighting this disease. Real people.
There it is.

Visit the Synovial Sarcoma Survivors Network at www.synovialsarcomasurvivors.org.

RESEARCH UPDATE

By Kevin B. Jones, MD, Principal Investigator, KB Jones Lab Research
Huntsman Cancer Institute, University of Utah

Synovial sarcoma is a cancer that impacts our society so much more than its numbers would suggest. While its victims may be counted in the hundreds, rather than the tens of thousands, most of these individuals develop synovial sarcoma during adolescence or young adulthood, when they are full of potential and promise for life.

Because we do not yet have a better strategy, we still treat synovial sarcomas with traditional chemotherapy treatments that are poisonous to all fast growing cells. These treatments try to slow the growth of the cancer cells by damaging their genetic code. A fast growing cell will often die when faced with this damage, rather than try to repair itself. However, a few cells will usually be given a paradoxical growth advantage by that damage. Synovial sarcoma, in particular, is a cancer that has very little gene damage at baseline. Therefore, treating it with agents that increase the gene damage, but fail to kill all the cancer cells, may actually be creating even more deadly versions of the same cancer that return after treatments are complete.

Our research efforts are therefore focused on trying to find other biology in synovial sarcoma to target with improved treatments, beyond merely damaging its genetic code. To do this, we are studying the specific cellular engines that synovial sarcoma cells co-opt to help themselves grow and spread. We are working on defining three of these specific engines in the lab right now. One is a cellular engine that regulates the expression of all the genes that give a cell the identity of a synovial sarcoma. Another is a cellular engine that controls the cell's response to stress signals. The third is a cellular engine that helps the cell respond to the very gene damage that so many of our traditional treatments cause. A fourth engine that we are beginning to dig into is the engine by which synovial sarcoma cells gain the capacity to spread through the body and threaten the life of their victim.

NEW PUBLICATIONS

Barrott JJ, Illum BE, Jin H, Hedberg ML, Wang Y, Grossmann A, Haldar M, Capecchi MR, Jones KB. Paracrine osteoprotegerin and β-catenin stabilization support synovial sarcomagenesis in periosteal cells. J Clin Invest. 2018 Jan 2;128(1):207-218. doi: 10.1172/JCI94955. Epub 2017 Nov 20. PubMed PMID: 29202462; PubMed Central PMCID: PMC5749548.

Banito A, Li X, Laporte AN, Roe JS, Sanchez-Vega F, Huang CH, Dancsok AR, Hatzi K, Chen CC, Tschaharganeh DF, Chandwani R, Tasdemir N, Jones KB, Capecchi MR, Vakoc CR, Schultz N, Ladanyi M, Nielsen TO, Lowe SW. The SS18-SSX Oncoprotein Hijacks KDM2B-PRC1.1 to Drive Synovial Sarcoma. Cancer Cell. 2018 Aug 13;34(2):346-348. doi: 10.1016/j.ccell.2018.07.006. PubMed PMID: 30107180.

Cancer Genome Atlas Research Network. Electronic address: elizabeth.demicco@sinaihealthsystem.ca; Cancer Genome Atlas Research Network. Comprehensive and Integrated Genomic Characterization of Adult Soft Tissue Sarcomas. Cell. 2017 Nov 2;171(4):950-965.e28. doi: 10.1016/j.cell.2017.10.014. PubMed PMID: 29100075; PubMed Central PMCID: PMC5693358.

HOW TO DONATE

Option 1: Write a check to *The Paul Nabil Bustany Fund for Synovial Sarcoma Research*. Send your check to 15 Footes Lane, Morristown, NJ, 07960.

Option 2: Donate online at www.pnbustanyfund.org.

The PNB Fund is a 501(c)(3) nonprofit organization. All donations are tax deductible.

The purpose of the Paul Nabil Bustany Fund for Synovial Sarcoma Research is to raise money for basic scientific research on synovial sarcoma. The PNB Fund is entirely run by volunteers and all donations go directly to synovial sarcoma research. Thank you for your support.